



## Case Report

## Sclerosing mediastinitis in a smoker with suspected lung cancer

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## ABSTRACT

A case history is presented of a middle-age patient with a rare cause of haemoptysis presenting as a pulmonary mass associated with mediastinal lymphadenopathy. Surgical resection demonstrated sclerosing (or fibrosing) mediastinitis, a condition uncommon outside areas of endemic fungal infection. We describe a case whereby an FDG-PET scan showed avid uptake of signal highly suspicious of cancer.

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## 1. History

A 44-year-old male smoker with a 36 pack year history of smoking presented acutely with a brisk haemoptysis. There was no history of pulmonary tuberculosis or a prior respiratory history. He did not have a cough, pleuritic chest pain, weight loss or night sweats, or history of domestic animal bites or exotic travel. Oxygenation in room air was normal and general and respiratory examination was unremarkable. Routine haematology, biochemistry, inflammatory markers and coagulation profile were similarly normal. The chest radiograph showed prominence of the right hilum with no evidence of a consolidation. A computed tomography (CT) scan showed a 2.5 cm mass lying medially in the right upper lobe, with a further mass immediately below this at the right hilum, measuring approximately 3 cm. Several small (<1 cm) peripheral pulmonary nodules were also noted, with minor enlargement of the right paratracheal and subcarinal nodes (see Fig. 1a,b).

A bronchoscopy showed mucosal distortion of the right upper lobe with no evidence of tumour macroscopically or on bronchial brush cytology. Microbiological culture of bronchial lavage was sterile including mycobacteria. Several weeks later, the haemoptysis had settled but the chest radiograph remained unchanged so he underwent a rigid bronchoscopy and

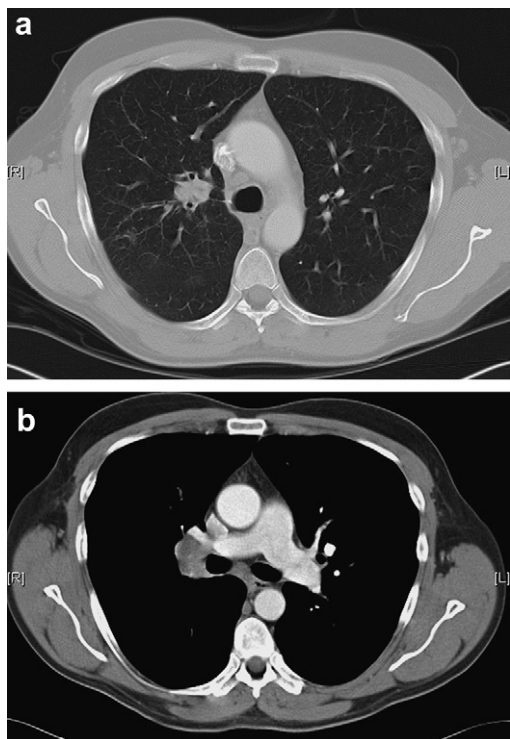
mediastinoscopy. Again, no new endobronchial abnormality was visible and there was now no evidence of tracheal lymphadenopathy. The patient remained well with no further haemoptyses over the next 3 months, at which stage a repeat CT scan showed no progression in the mass or mediastinal lymphadenopathy and, as EBUS was not available locally, a further repeat bronchoscopy was performed, confirming mucosal erythema and distortion. Further multiple bronchial biopsies showed no evidence of neoplasia and bronchial lavage was again sterile on microbiological culture. A PET (Positron Emission Tomography) CT scan was performed using fluorine-18 fluorodeoxyglucose (F-18 FDG). This demonstrated significantly increased activity at the right hilum congruent with the right hilar mass, but the mass had not increased significantly in size from the earlier CT scan (Fig. 2). As the patient had remained stable, he did not wish to consider a further thoracoscopy or thoracotomy.

Three months later he re-presented with further episodes of haemoptysis and so he agreed to proceed to a right thoracotomy and resection. The lesion was not fully resectable, and he continued to have significant bleeding and haemoptyses in the operative period. Control of bleeding was not possible without proceeding to a right pneumonectomy. This controlled both his symptoms, bleeding and he had no further haemoptyses.

Histological examination of the resected mass and lymph nodes showed infiltration by dense hyalinised fibrous tissue with chronic inflammation and giant cells. There was no evidence of malignancy within the mass or the lymph nodes. The features were consistent with sclerosing mediastinitis (see Fig. 3). Subsequent serological examination did not show evidence of fungal infection.

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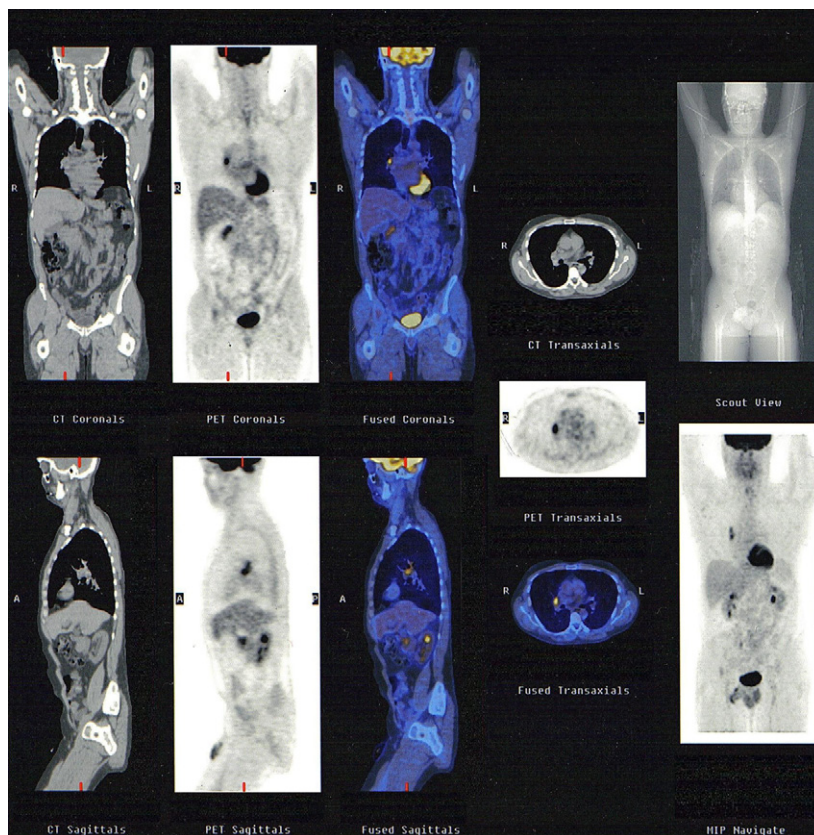


**Fig. 1.** (a) and (b): Lung and mediastinal windows respectively, from a staging-protocol CT scan.

## 2. Discussion

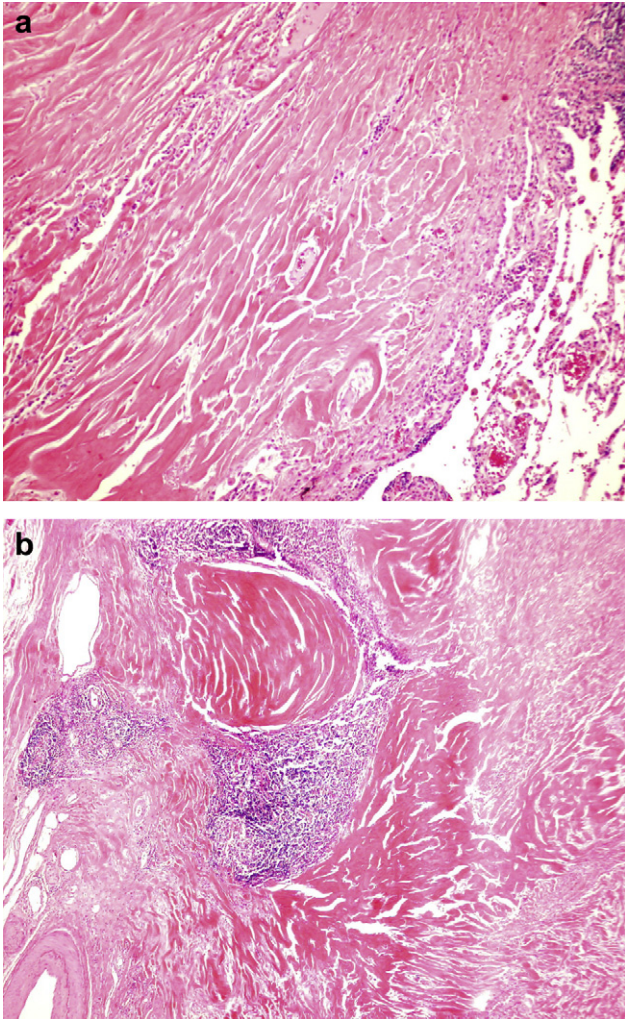
Sclerosing (or fibrosing) mediastinitis is a rare but benign disorder characterized by inflammation and dense fibrosis of mediastinal structures, and is associated with a variety of clinical syndromes. The cause is probably an abnormal fibroproliferative response to an inflammatory stimulus, most commonly seen after a granulomatous infection secondary to the fungus *Histoplasma capsulatum*. It is also associated with other idiopathic fibrotic diseases, such as sclerosing cholangitis, Reidel's thyroiditis and retroperitoneal fibrosis. Rarely, it has been described in association with autoimmune disease, Behcet's disease, tuberculosis, malignancy and after radiation therapy.<sup>1–3</sup> The inflammatory stimulus leads to encasement of mediastinal structures within a dense fibrotic mass, arising from an invasive chronic inflammatory process causing erosion as well as external compression of structures such as the superior vena cava, pulmonary veins or arteries, central airways, or oesophagus, leading to different clinical symptoms such as dyspnoea, dysphagia or haemoptysis, or on occasion is asymptomatic.<sup>3</sup> It can masquerade as thoracic cancer in countries outside endemic fungal infection areas, including Europe.

A spectrum of focal and diffuse fibrosing mediastinitis is recognised; diffuse mediastinitis presents as an infiltrating, often non-calcified mass that affects multiple mediastinal compartments while the localized form leads to a calcified mass in the paratracheal or subcarinal areas of the mediastinum or in the pulmonary hila. In milder forms of the disorder the chest X-ray can be unremarkable. In progressive disease, both computed tomographic (CT) or magnetic resonance (MR) imaging can define the extent of disease although



**Fig. 2.** The CT image shows soft tissue thickening around the right hilum compatible with enlarged lymph nodes, and the PET image shows a 'hot' spot of avid FDG uptake in the same area.





**Fig. 3.** (a) and (b) respectively show a haematoxylin and eosin-stained high power photomicrograph of dense hyalinised fibrous tissue with chronic inflammation and adjacent lung parenchyma (3a) and the dense collagenous tissue extending into a hilar node and surrounding airways and vascular channels at the hilum (3b).

distinguishing from a malignant process can be difficult despite the imaging modalities.<sup>4</sup>

The diagnosis of sclerosing mediastinitis in endemic areas involves identifying *Histoplasma* by microbial culture (but this is costly and time-consuming) or by serological response, but even this approach is associated with false-positive results in individuals from endemic areas without the disorder, and false negatives

results in those with compromised immune function. A positive delayed hypersensitivity skin test also cannot discriminate between past or current infection. As the disorder is associated with other clinical entities in the absence of *Histoplasma* infection, a surgical biopsy is usually required. Consequently the oral antifungal agent ketoconazole is useful in patients with proven *Histoplasma* infection,<sup>5</sup> but often as an adjunct to surgery which is required initially for diagnosis, to rule out cancer and to treat the complications. The extent of invasion of mediastinal structures determines surgical success, and procedures such as superior vena caval reconstruction, tracheal or oesophageal decompression, lobectomy, division of tracheo-oesophageal fistulae or rarely a pneumonectomy are required.<sup>6</sup> Oral corticosteroid therapy has been proven to be useful in selected cases.<sup>7,8</sup>

### 3. Conclusion

Sclerosing mediastinitis is rare in Europe but should be considered in patients with a lung or mediastinal mass of unknown aetiology. Surgical intervention is often required to make the diagnosis and, in some cases to control symptoms and treat complications. Medical treatment should also be considered, particularly if there is confirmed *Histoplasmosis*. The long-term prognosis is variable depending on the extent of mediastinal involvement and outcome of surgical treatment of complications. Overall it is good if not associated with a malignancy.

### Competing interests

None declared.

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